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CLINICAL REPORT

Recurrent Legg-Perthes-Calvé disease

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KEYWORDS

Legg-Perthes-Calvé disease;
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Summary

Introduction: Legg-Perthes-Calvé disease (LPC), or primitive hip osteochondritis, is a frequent pathology but in which recurrence is extremely rare.

We report: The case of a girl diagnosed with bilateral LPC at the age of 4.5 years. Containment in a Scottish-Rite (Atlanta) brace was prescribed. X-ray follow-up found complete right-hip healing at 1.5 years' evolution (Catterall group 2). The patient was seen again at the age of 8 years, for limp and functional disorder associated with recurrence of right-hip LPC (Catterall group 3). Evolution was satisfactory: the girl was assessed at the time of bone maturity, with good clinical and radiological findings.

Discussion: Children contracting LPC are commonly thought to be thereby "vaccinated" against it. This is wrong, and a literature search found 10 cases similar to the present one, making 11 children in all (two girls, nine boys) presenting with recurrent LPC. Mean age at initial onset was 4 years (range, 2.5–6 yrs). Five of the 11 had initially been presented with bilateral LPC. Mean age at recurrence was 9.4 years (range, 6–12 yrs). The recurrences were more severe than the initial episodes, but final prognosis after recurrence would not seem to be worse than normal.

Conclusion: This exceptional case of recurrent LPC was well documented up to bone maturity. It does not support the notion of Meyer's disease at the initial episode, as suggested by certain authors, but rather that of true recurrence of the primitive LPC.

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Introduction

Legg-Perthes-Calvé disease (LPC), or primitive osteochondritis of the hip, is a frequent pathology-causing limp in

children aged 3–9 years. Victims are commonly thought to be as they were "immunized" against recurrence. Only a few cases of recurrence have in fact been reported [1–9]; severity and evolution at bone maturity, however, were seldom specified.

In 40 years' experience in pediatric orthopedics, a single case of recurrent LPC has been managed in our department. The present case analysis and literature review is intended to improve understanding of this disease entity.

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Figure 1 Various evolutive phases of osteochondritis on frontal pelvic X-ray. Right (overlying) and left (underlying) hip views for the same period are superimposed. The right femoral epiphysis is reconstructed on the October 1999 view, while the left remains fragmented.

Clinical case

Anaïs was a 4 1/2 year-old girl, free of medico-surgical history, when bilateral LPC was diagnosed in December 1997. She had previously exhibited two consecutive episodes of limping with pain in both hips. Bilateral involvement was not simultaneous, as can be seen from an older X-ray view (January 1996), showing initial left involvement with a normal right hip at that point in time. Fig. 1 shows the radiologic evolution of this first period.

The girl was referred to our department in May 1998 for bilateral LPC; she had then been managed by means of a Scottish-Rite (Atlanta) brace for 1 year. The right hip was classified as Catterall group 2 [10] and Herring group A [11], and the left as respectively group 3 and A.

In July 1998, the patient was pain-free, with good hip-joint amplitudes (abduction 50°, adduction 30°, internal rotation 50° in the right hip, and abduction 50°, adduction 30°, internal rotation 40° in the left), and the brace was removed.

When seen again in February 1999, she was symptom-free, with both hips normal on clinical assessment. X-ray found the right femoral head in reossification (Fig. 1). Anaïs was allowed to resume normal life, without sports restriction, and referred to her local orthopedic physician for follow-up. Radiography performed in October 1999 (Fig. 1) found the right femoral epiphysis reconstructed.

The patient was again referred to us by her pediatrician in September 2001, for recurrence of right hip pain of a few months' evolution. Fig. 2 shows the radiologic evolution of this episode. Forearm crutches had been prescribed in view of the degree of pain. Joint amplitude was at that point 0-120-10-20-20-10 in the right hip and 0-140-40-20-60-30 in the left. Right-hip X-ray performed in August 2001 found a characteristic radiolucent crescent line.

Given the exceptional nature of recurrence in LPC, inflammation assessment with MRI and bone-scan were performed; the former proved normal, whereas the bone-scan (Fig. 3) and MRI (Fig. 4) confirmed the diagnosis of right-hip recurrence of LPC.

Examination for secondary LPC proved normal (blood count, CRP, and C protein, S protein, antithrombin 3 and D-dimer assays). Testing for antiphospholipid syndrome, activated C-protein resistance, factor-V Leiden mutation and prothrombin gene mutation were all negative.

Knee X-ray, performed at the time of the initial episode to rule out multiple epiphyseal dysplasia was normal. The patient was managed by nocturnal traction and daytime Thomas' hip splint. Clinical and radiological evolution were classical, although more severe than in the initial episode: Catterall group 3 [10] and Herring group A [11].

In March 2002, her hips were free of pain and with mobility improved by the traction (0-120-30-20-30-20 in the right hip and 0-120-45-20-50-40 in the left). Radiography found right osteochondritis in reossification phase with conserved lateral pillar (Fig. 2). Nocturnal traction was stopped in June 2002.

In March 2003, the Thomas' hip splint was progressively replaced by forearm crutches. Clinical examination found 10mm right thigh amyotrophy, 10° symmetric anteversion and good hip mobility. X-ray found the femoral epiphysis in reossification. Sports were resumed in June 2004.

Assessment in May 2006 found occasional pain following prolonged sport. Examination found symmetrical hip-joint amplitude and no thigh amyotrophy. X-ray found good femoral head remodeling with right coxa magna but good sphericity.

At last follow-up (March 2009), at the age of 15 years, the patient was totally symptom-free. Joint amplitudes were symmetrical. X-ray showed only slight sequelae (Fig. 5).

Table 1 Cases of recurrent Legg-Perthes-Calvé disease in the literature (radiologic cure of 1st episode was claimed in all cases).

Author/date	Gender	Age at 1st episode (yrs)	Side of 1st episode	Catterall group 1st episode	Age at recurrence	Side of recurrence	Catterall group recurrence
Kemp et al. [1] 1971	M	4	R	1	7.5	R	2
Katz [2] 1973	M	5.5	R + L	ns	11	R	ns
Bjerkreim and Hauge [3] 1976 case No.1	M	3	L	2	12	L	3
Bjerkreim and Hauge [3] 1976 case No.2	M	2.5	R	2	8	R	3
Axer and Hendel [4] 1977	M	4	L	ns	8	L	ns
Burkhead and Wenger [5] 1981	M	6	R + L	2	10	L	3
Martinez and Weinstein [6] 1991	F	3	R + L	1	10	R	2 then 4*
Schonecker et al. [7] 1998	M	3	R	2	6	R	4
Stevens et al. [8] 2001	M	5.5	L	1	11	L	2
Ghanem et al. [9] 2005	M	2.5	R + L	2	11.5	L	3
Present case	F	4.5	R + L	2	8	R	3
Total/mean	9M 2F	4 yrs	5B + 6U	1 or 2	9.4 yrs	6R-5L	2-3 or 4

F: female; M: male; ns: not specified; B: bilateral; U: unilateral.

* Change of Catterall group over 2nd episode evolution.

Perthes disease'', and hypothesized that cases of recurrent osteochondritis, including their own two (which had, however, been originally unilateral) were cases of Meyer's disease [13] subsequently developing into true osteochondritis. We dispute this attitude: in the present case, the right hip involvement may be open to discussion, but that of the left hip displayed all the features of the severe form described by Legg, Perthes and Calvé. That one and the same patient should present with unilateral Meyer's dysplasia [13] and contralateral primitive osteochondritis appears improbable to say the least.



Figure 5 Frontal pelvic X-ray at last FU, March 2009 (13 years after initial osteochondritis).

In all the cases of bilateral osteochondritis except that reported by Katz [2], the side showing recurrence was the one with the faster and more benign initial evolution. All these bilateral involvements had been managed by Scottish-Rite braces.

In the published cases and in the present, recurrence was more severe than the initial episode (Table 1). This may be a bias induced by greater age at recurrence: the later the onset, the poorer the prognosis (Ghanem et al. [9]).

Reports of recurrent LPC generally focus on diagnosis and the circumstances surrounding recurrence (Bjerkreim and Hauge [3], Axer and Hendel [4], Katz [2], Ghanem et al. [9]), with few data as to evolution at bone maturity.

The cases reported by Burkhead and Wenger [5] and by Schonecker [7] showed unfavorable evolution, requiring surgery. In that reported by Martinez and Weinstein [6], surgery was considered for hip stiffness and femoral head deformity at last follow-up. Finally, Stevens' [8] patient was pain-free and with very good hip mobility at last follow-up, but with a 2.5 cm difference in femoral length due to a shortened neck, and congruent coxa magna.

Conclusion

Recurrent LPC is possible but exceptional (one case out of 700 in our own department). It mainly affects boys presenting mild initial osteochondritis before the age of 6 years. The description and evolution of the present extra case do not argue in favor of the hypothesis of initial Meyer's disease [13] made by certain authors.

The evolution of the hip showing recurrence tends to be more severe than in the initial episode, but is not necessarily of poor prognosis.

Conflict of interest statement

No conflict of interest for any of the authors.

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